Indian J Lepr 2019, 91 : 159-164

http://www.ijl.org.in

Case Report

Hansen's Disease – Clinically Atypical Presentation with Atypical Histopathology

Vinitha G[1], Asha T[2], TP Thankappan[3], Sulochana G[4]

Received : 21.06.2018 Accepted : 28.12.2019

Type II Lepra reaction is a Th2 mediated hypersensitivity reaction characterized by involvement of skin and nerves. Herein we report an unusual leprosy case that had a rheumatic mode of onset of Type II Lepra reaction. This case also had livedoid plaques, ulcers on the genitalia, extremities as the initial manifestation of Hansen's Disease and atypical histopathology. His symptoms had started 1 year back as polyarthralgia of both knees and small joints of hands and feet for which he was on immunosuppressant by a Rheumatologist who made a diagnosis of rheumatoid arthritis. After 6 months, patient developed red painful lesions; initially on arms, chest and trunk followed by legs. Subsequently, painful ulcerations developed on feet, knees and genitalia and a dermatology referral was made. Histopathology and slit skin smears led to diagnosis of Lepromatous leprosy in Type II Lepra reaction. Hansen's disease is a great imitator. Rheumatic manifestations can be the initial presentation of Type II Lepra reaction. Rheumatic manifestations thus can be the initial presentation of Type II Lepra reaction even before starting Multi drug therapy. High index of suspicion and routine slit skin smears in small peripheral settings can be helpful in diagnosing and treating such cases.

Key words : Type II Lepra Reaction, Rheumatic Onset, Leg and Genital Ulcers, Large Vacuolated Histiocytes

Introduction

Hansen's disease is a chronic infectious disease with predominant involvement of skin and peripheral nerves. Type II Lepra reaction, classically presents as erythematous nodules on the face, trunk and extremities (El Gandy et al 2016), however, it may have varied manifestations (Vijendran et al 2014). The rheumatic features, especially arthritis forms the third manifestation of the disease whose prevalence ranges from 60–80% in rheumatology clinics to 1 - 2% in dermatology clinics (Gupta et al 2016). The various histological features noted in Lepra reactions are neutrophilic infiltration, dermal oedema, vasculitis and panniculitis (Negera et al 2017). Herein we report an unusual case of Type II

[1] Dr Vinitha Gopalakrishnan, Consultant Dermatologist,
[2] Dr Asha Thankappan, Assistant Professor
[3] Dr Thonduparambil Pappan Thankappan, Professor
[4] Dr Sulochana Gopalan, Professor of Pathology
Pushpagiri Institute of Medical Sciences, Tiruvalla - 689101, Kerala, India
Corresponding author: Dr Vinitha Gopalakrishnan, VPS Lakeshore Hospital and Research Centre, Nettoor, Kochi, Kerala, India,
Email : drvinitha@gmail.com

Lepra reaction with rheumatic manifestations, livedoid lesions, genital ulceration and atypical histopathology.

Case Report

A 68 year old male presented to our Dermatology Outpatient Department with oozing and extensive ulceration of both feet of one-month duration. The symptoms started 1 year back as polyarthralgia of both knees and small joints of hands and feet for which he was on immunosuppressant by a Rheumatologist who made a diagnosis of rheumatoid arthritis. After 6 months, patient developed red painful lesions; initially on arms, chest and trunk followed by legs. Over a period of 2 weeks, painful ulcerations developed on feet, knees and genitalia and a dermatology referral was made. On clinical examination, there were multiple dull erythematous livedoid plaques of varying sizes distributed over forearms (Fig. 1a), thighs, legs (Fig. 1b) trunk and abdomen (Fig. 1c). Erosion and ulcerations were noted over multiple toe digits, sole of left foot (Fig. 1d), right knee and tip of penis. No nodules, necrosis or peripheral nerve thickening. Face, oral mucosa, hair and nails were normal. Focal sensory loss over both feet was noted. Routine blood examination revealed an elevated ESR of 130 mm/hr, deranged renal function (Blood urea: 50 mg/dl: S.

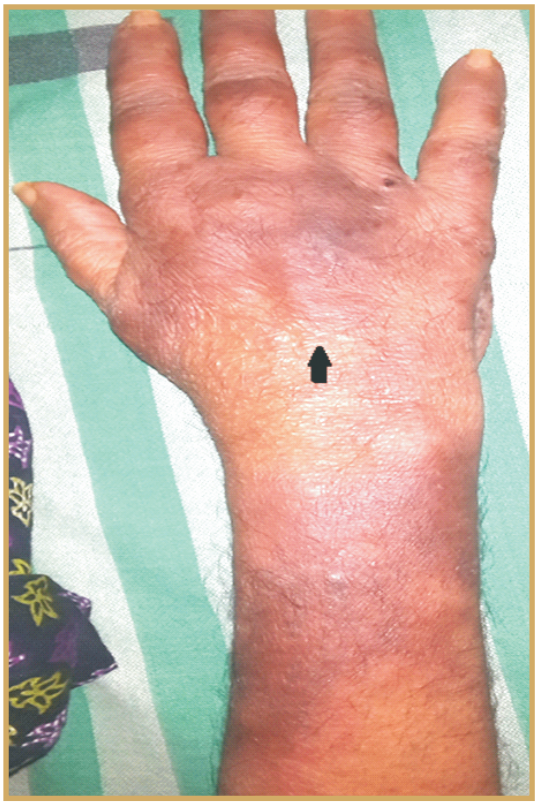

Fig 1a : Dull erythematous plaques over forearms

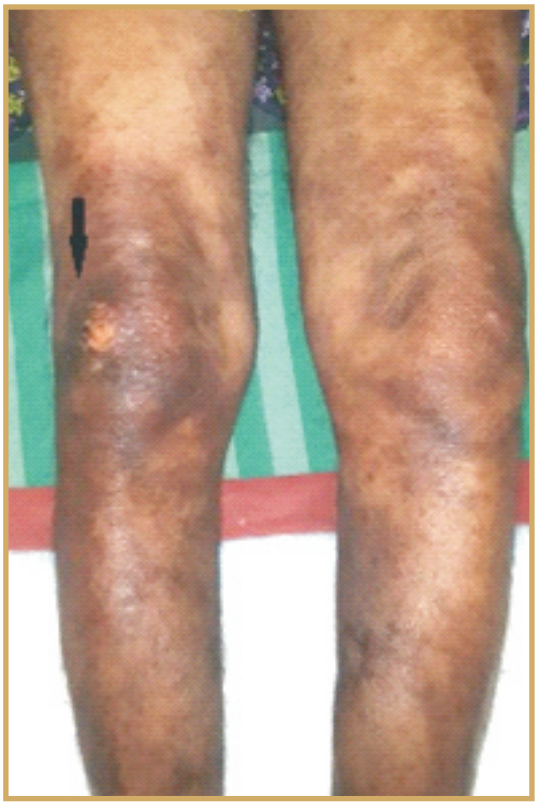

Fig 1b : Livedoid plaques over legs

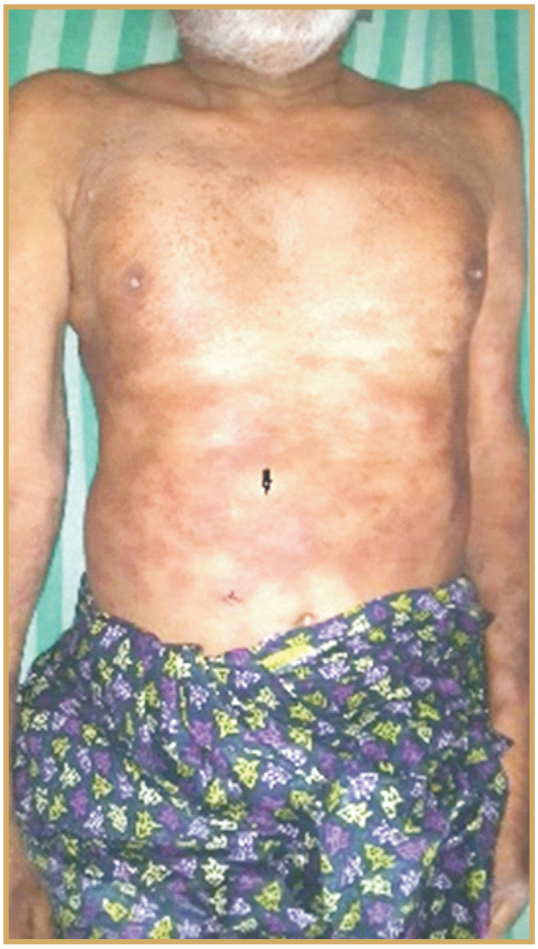

Fig 1c : Dusky red plaques of varying size on the abdomen

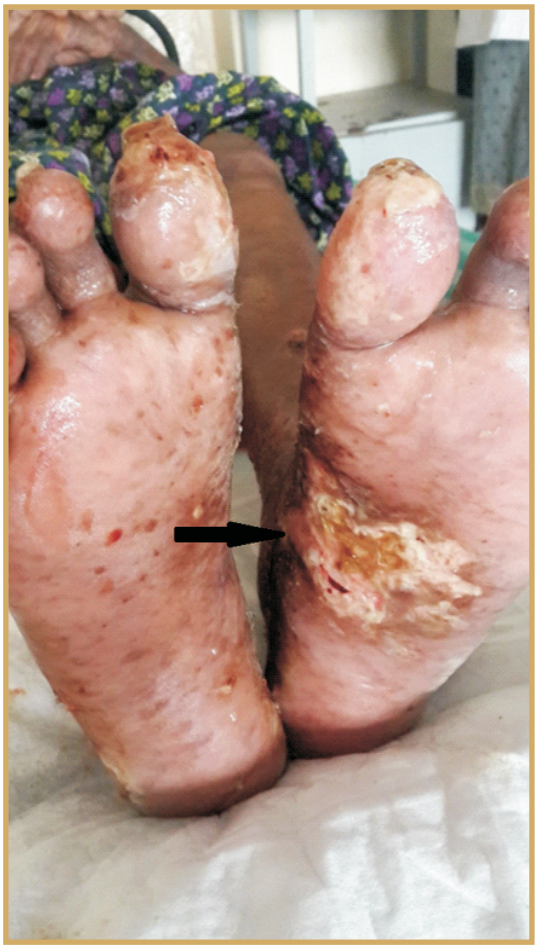

Fig 1d : Ulceration sole of foot

Creatinine: 1.61 mg/dl), altered 24 hour urine protein (480 mg / day) and positive RA factor. Autoimmune profile, Chest Xray, USG Abdomen, CT Chest were noncontributory. A Tzanck smear from the ulcer revealed inflammatory cells. Considering differential diagnosis of Hansen's Disease and Systemic Necrotizing Vasculitis, biopsy was taken from abdomen and earlobe.

Slit skin smears from ear lobe (Fig. 2a) showed AFB positive rods with a Bacillary Index 6 (+).

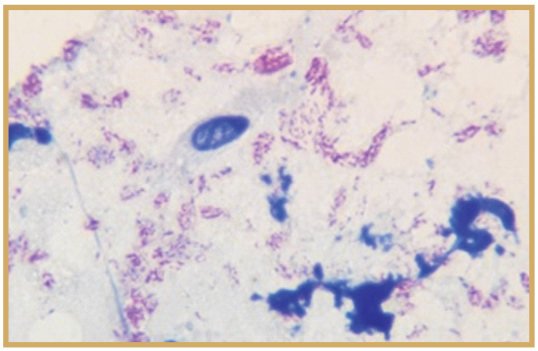

Fig 2a : Modified Ziehl-Neelsen stain BI 6 (+)

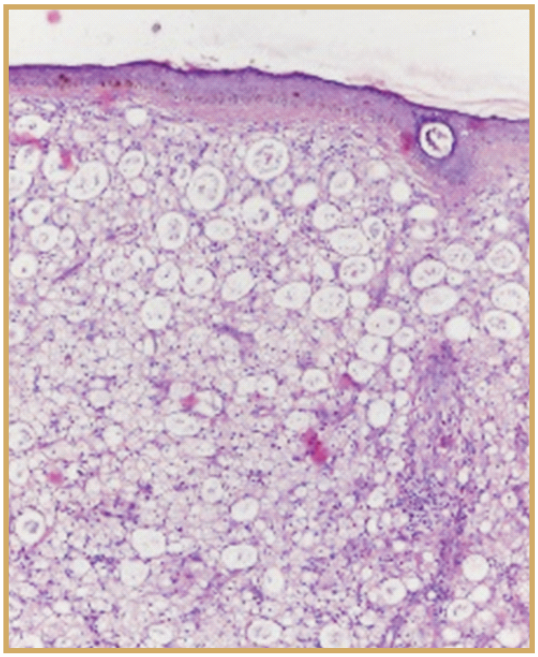

Fig 2b : Ear lobe (H&E 10X), Flattened epidermis with grenz zone and sheets of large vacuolated histiocytes

On histopathological examination, ear lobe biopsy showed flattened epidermis with a Grenz zone; below it were sheets of foamy histiocytes and scattered lymphocytes (Fig. 2b) and specimen from the abdomen revealed dermis showing small vessel vasculitis with scanty infiltrate of lymphocytes and neutrophils (Fig. 2c). Among the histiocytes, large vacuolated cells with eosinophilic fragmented granulated material were seen (Fig. 2d). On Fite Faraco stain of both the slides, foam cells loaded with bundles of cigar shaped lepra bacilli were seen. (Fig. 2e). So considering the clinical features and histopathology, a diagnosis of Lepromatous leprosy in Type II Lepra reaction with rheumatic mode of onset was made and the patient was started on Multi Drug Therapy.

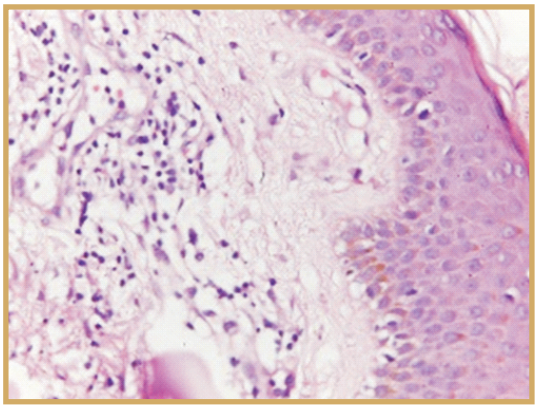

Fig 2c : Abdomen (H&E 40x) : small vessel vasculitis with inflammatory infiltrate

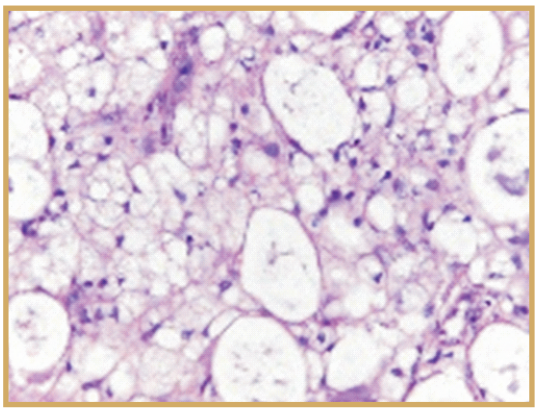

Fig 2d : Abdomen (H&E 40x), Foamy histiocytes with eosinophilic granulated materials

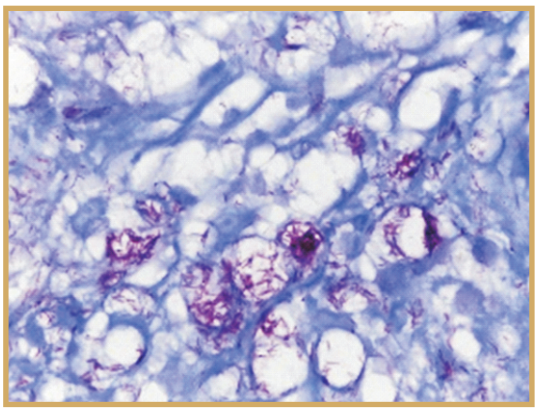

Fig 2e : Abdomen (Fite Faraco stain. 40x), showing foam cells with bacilli

Table 1 : Rheumatic manifestations of leprosy (El-Gendy et al 2016, Gupta et al 2016)

S.No.	Major Category	Clinical presentation
1.	Leprosy related arthritis	i. Symmetric inflammatory polyarthritis
		ii. Rheumatoid arthritis like
		iii. Neuropathic type (Charcots joints)
		iv. Septic arthritis
		v. Polyarthralgias
2.	Types of vasculitis like presentation reported in patients with Hansen's disease	i. Leukocytoclastic vasculitis
		ii. Polyarteritis nodosa like
		iii. Necrotising cutaneous and systemic vasculitis
		iv. Purpura fulminans
		v. Cryoglobulinemia
		vi. Digital vasculitis
		vii. Gangrene
3.	Connective Tissue Diseases like presentation reported in Hansen's disease	i. SLE like presentation of Hansen's disease
		ii. Dermatomyositis - like features
		iii. Seronegative symmetric synovitis as feature of Hansen's disease
4.	Autoantibodies reported in sera of patients with Hansen's disease	i. RF (most common)
		ii. Antinuclear antibody (ANA - speckled pattern)
		iii. Anti-SS-B
		iv. Antimitochondrial (AMA)
		v. Antithyroglobulin antibodies.
		vi. Antineutrophil cytoplasmic antibodies (p-ANCA >> c-ANCA)

Discussion

Leprosy is a multifaceted disease as the clinical presentation is diverse and can mimic a wide variety of other diseases (Rath et al 2014). Rheumatic mode of onset of Hansen's disease has also been described and they form 1-2% (El-Gendy et al 2016, Gupta et al 2016). The various rheumatic manifestations of leprosy are summarized in Table 1.

The hall mark of type II reaction is Erythema Nodosum Leprosum. Lucio phenomenon; a rare form presents with tender nodules, ulceration, bulla formation, and necrotic lesions. Other manifestations of type II reactions include ulcers (peripheral neuropathy or Lucio phenomenon associated) neuritis, iritis, iridocyclitis, orchitis, glomerulonephritis, arthritis, and lymphadenopathy. Type II reaction mainly occur after initiation of Multidrug therapy in 90% cases; rarely it can be the initial manifestation of the disease too (Chauhan et al 2006).

Our case was misinterpreted for a vasculopathy or a connective tissue disease because of the multisystem involvement (anemia, proteinuria,

renal impairment, hypothyroidism, peripheral neuropathy, and polyarthralgia and pedal oedema). Though there are many reports and reviews of polyarthritis as initial presentation of Hansen's (Chauhan et al 2010, Prasad et al 2013, Saraswat et al 2019), generalized livedoid lesions, ulcers of extremities, knees and genitalia as initial manifestations of the disease as in our case is very rare.

Typical histology of type II Lepra reaction characterized by dermal edema, neutrophilic infiltration with or without vasculitis on a background of macrophage granuloma (Adhe et al 2012). In our case lesions from abdomen showed small vessel vasculitis with scanty neutrophils. No macrophage granulomas were noted. Dermis showed foamy histiocytes among which were large vacuolated cells with eosinophilic fragmented granulated material. The large sized histiocytes with fragmented granulations are not seen in classical cases of lepromatous leprosy. It may be attributed to the chronic immunosuppression and heavy bacillary load.

Conclusion

Leprosy has wide variety of clinical presentations. Rheumatic mode of onset of type II Lepra reaction may be the initial manifestation of Hansen's disease per se. High index of suspicion is essential for arriving at correct diagnosis of leprosy presenting with atypical presentations which can even be made with slit skin smears in peripheral settings.

References

1. Adhe V, Dongre A, Khopkar U (2012). A retrospective analysis of histopathology of 64 cases of lepra reactions*. Indian J Dermatol*. **57**: 114-7.
2. Chauhan S, D'Cruz S, Mohan H et al (2006). Type II lepra reaction: an unusual presentation. *Dermatol Online J.* **12(1**): 18. PubMed PMID: 16638386.
3. Chauhan S, Wakhlu A, Agarwal V (2010). Arthritis in leprosy. *Rheumatology*. **49**: 2237-2242.
4. El-Gendy H, El-Gohary RM, Shohdy KS et al (2016). Leprosy Masquerading as Systemic Rheumatic Diseases. *J Clin Rheumatol*. **22**: 264-71.
5. Gupta L, Zanwar A, Wakhlu A et al (2016). Leprosy in the rheumatology clinic: an update on this great mimic. *Int J Rheum Dis.* **19**: 941-945.
6. Jopling WH, McDougall AC (1996). Handbook of leprosy, 5th edition, Delhi, CBS Publishers and Distributors, 10-53.
7. Negera E, Walker SL, Girma S et al (2017). Clinico-pathological features of erythema nodosum leprosum: A case - control study at ALERT hospital, Ethiopia. *PLoS Negl Trop Dis.* **11(10):** e0006011.
8. Prasad S, Mishra R, Aggarwal A et al (2013). Leprosy revealed in a rheumatology clinic: a case series. *Int J Rheum Dis*. **16**: 129-133.
9. Rath D, Bhargava S, Kundu BK (2014). Leprosy mimicking common rheumatologic entities: a trial for the clinician in the era of biologics. *Case Rep Rheumatol*. 2014:429698. doi: 10.1155/2014/429698. Epub 2014 Nov 6. PubMed PMID: 25431725; PubMed.
10. Saraswat N, Agarwal R, Chopra A et al (2019). Interesting and Unusual Presentation(s) of Leprosy Resulting in Delayed diagnosis. *Indian J Lepr*. **91**: 47-54.
11. Vijendran P, Verma R, Vasudevan B et al (2014). Rare atypical presentations in Type 2 lepra reaction: a case series. *Int J Dermatol.* **53**: 323-6.

How to cite this article : Vinitha G, Asha T, Thankappan TP et al (2019). Hansen's Disease – Clinically Atypical Presentation with Atypical Histopathology. *Indian J Lepr.* **91**: 159-164.